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Foix-Chavany-Marie syndrome due to unilateral anterior opercular infarction with leukoaraiosis

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ABSTRACT

Foix-Chavany-Marie syndrome (FCMS) is a cortical-subcortical pseudobulbar palsy characterized by automatic voluntary dissociation of facio-masticatory-pharyngo-glosso-laryngeal movements. FCMS is typically caused by vascular insults on the bilateral anterior opercular or adjacent subcortical areas. Acute onset of FCMS secondary to a unilateral lesion is extremely rare. Herein we present a case of FCMS caused by acute unilateral anterior opercular infarction with preexisting bilateral leukoaraiosis. Our case shows that an acute unilateral anterior opercular lesion can decompensate preexisting corticobulbar-subcortical lesions and cause the typical features of FCMS.

KEYWORDS Anterior operculum; automatic voluntary dissociation; Foix-Chavany-Marie syndrome; pseudobulbar palsy; stroke

Foix-Chavany Marie Syndrome (FCMS) is a rare type of bilateral cortical-subcortical pseudobulbar palsy characterized by voluntary lingual, facial, pharyngeal, and masticatory paralysis with preservation of automatic and involuntary movements.[1–4] Patients usually present with facial weakness, severe dysphagia, spastic dysarthria, and anarthria on voluntary movements.[2] FCMS is typically caused by vascular insults on the bilateral anterior opercular or adjacent subcortical areas,[1–3] and acute onset secondary to a unilateral lesion is particularly rare. Here we describe a patient with preexisting bilateral leukoaraiosis who presented with acute onset of FCMS secondary to a unilateral anterior opercular infarction.

CASE REPORT

An 83-year-old right-handed Hispanic woman had a past medical history of hypertension, diabetes mellitus type 2, dyslipidemia, and ischemic stroke on the right frontal lobe without prior residual neurological deficit. She presented to the emergency department with an abrupt onset of severe slurred speech, trouble swallowing, facial weakness, and left-sided hemiparesis. On the initial neurological examination, she had severe spastic dysarthria, dysphagia, an upper motor neuron pattern of left facial paralysis, and left-sided hemiparesis (Medical Research Council motor strength 4/5, hyperreflexia 3+, and a positive Babinski sign on the left foot). Her mouth was half open and she could not close it when asked to do so, although she could close it spontaneously and involuntarily when she smiled. Sneezing and yawning were intact. She did not have sensory deficits, aphasia, alexia, or agraphia. The complete blood count and comprehensive metabolic panel were unremarkable. Brain magnetic resonance imaging (MRI) showed an area of restricted diffusion consistent with an acute infarction on the right frontal operculum without acute lesion on the brainstem or cerebellum. MRI also revealed encephalomalacia on the right frontal lobe secondary to chronic infarction and extensive leukoaraiosis on the bilateral subcortical white matter *(Figure 1)*. Video fluoroscopic swallow revealed a single episode of penetration on the last swallow of thin liquids with no aspiration, and free spill was noted, which confirmed oropharyngeal dysphagia. At 2-week follow-up, her speech, swallowing, and voluntary facial movements showed partial improvement.

DISCUSSION

The current mapping of the divisions of the operculum has demonstrated that stimulation causes motor and language deficits as well as somatosensory and oropharyngeal symptoms.[5] The anterior operculum contains the voluntary motor fibers for the

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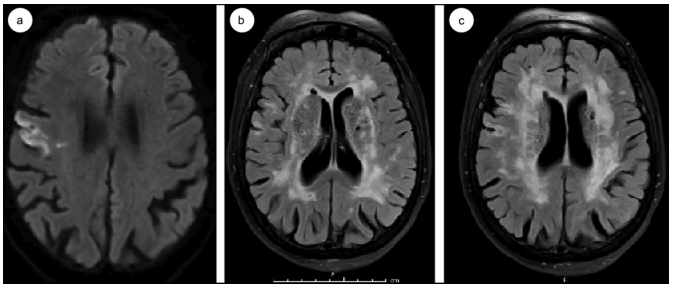

Figure 1. Patient MRI showing **(a)** acute infarction of the right frontal operculum and **(b, c)** encephalomalacia on the right frontal lobe and extensive leukoaraiosis on the bilateral subcortical white matter.

5th, 7th, 9th, 10th, and 12th cranial nerves, which then travel to the cranial nerve nuclei via the corticobulbar tract.[1,2] Injury to this area can cause FCMS bilateral voluntary paralysis of facial, masticatory, pharyngeal, laryngeal, and brachial muscles.[6–8] Autonomic-voluntary dissociation in FCMS is explained by the presence of alternative pathways for facial emotional expression and automatic movements, hypothesized to be mediated through the inner forebrain and outer longitudinal bundle that connect the amygdala and hypothalamus to the brainstem.[1,2]

A unilateral lesion can cause FCMS either independently or if a patient has experienced contralateral cortical-subcortical lesions prior to the recent injury.[1–3,8] Magnus et al reported the first case in 1837,[9] and in 1988 Starkstein et al[10] reported a patient with a lesion in the right insula and frontotemporoparietal operculum. However, these cases were reported before the advent of MRI or nuclear scanning. We found three fairly recent cases of an isolated unilateral lesion resulting in FCMS; each report described a patient who developed FCMS after unilateral damage to the pars opercularis, two due to a stroke and one due to iatrogenic surgical error.[11–13] Previous case reports have described how a new unilateral lesion in combination with older lesions can result in FCMS.[14,15] Our patient's brain MRI revealed some previous brain damage contralateral to the acute infarction, which appears to have contributed to FCMS. Some hypotheses as to why a contralateral lesion causes FCMS include (1) contralateral subcortical lesions interrupt the corticobulbar projections from the anterior opercular cortex to the brainstem nuclei, and (2) a unilateral representation of the motor centers dominantly and bilaterally innervates the affected muscles.[1] Martino et al used tractography to correlate the occurrence of FCMS to the resection of connections between the frontal aslant tract and arcuate fasciculus and the right pars opercularis for the first time in the literature.[12] A case of unilateral FCMS with right infarction of the corona radiata revealed decreased volume of bilateral cortico-nuclear tract fibers in tractography; when left cortico-nuclear damage was added, it resulted in bilateral disability.[2]

To our knowledge, FCMS secondary to an acute unilateral opercular lesion with underlying extensive leukoaraiosis has not been reported. In our case, we propose that the corticobulbar tract may have already been affected by the chronic white matter lesions, but these slowly accumulated over time, which allowed the patient to asymptomatically compensate for this damage. These lesions, in addition to the acute opercular infarction, may have resulted in the bilateral corticobulbar dysfunction manifesting as FCMS. Prognosis for unilateral opercular lesions tends to be better than for bilateral lesions.[16] Our patient partially recovered within a couple of weeks. Since subcortical leukoaraiosis is not uncommon, particularly in aging populations, FCMS due to an acute unilateral lesion should be considered in patients presenting with acute onset of bilateral pseudobulbar palsy.

1. Sá F, Cordeiro IM, Mestre S, Nzwalo H. Unilateral opercular infarction presenting with Foix-Chavany-Marie syndrome. *Case Rep*. 2014; 2014:bcr2014206439. doi:10.1136/bcr-2014-206439.
2. Yoshii F, Sugiyama H, Kodama K, Irino T. Foix-Chavany-Marie syndrome due to unilateral anterior opercular damage with contralateral infarction of corona radiata. *Case Rep Neurol*. 2019;11(3):319–324. doi: 10.1159/000503856.
3. Ohtomo R, Iwata A, Tsuji S. Unilateral opercular infarction presenting with Foix-Chavany-Marie syndrome. *J Stroke Cerebrovasc Dis*. 2014;23(1):179–181. doi:10.1016/j.jstrokecerebrovasdis.2012.08.015.
4. Nitta N, Shiino A, Sakaue Y, Nozaki K. Foix-Chavany-Marie syndrome after unilateral anterior opercular contusion: a case report. *Clin Neurol Neurosurg*. 2013;115(8):1539–1541. doi:10.1016/j.clineuro.2012.12.036.
5. Mălîia M-D, Donos C, Barborica A, et al. Functional mapping and effective connectivity of the human operculum. *Cortex*. 2018;109: 303–321. doi:10.1016/j.cortex.2018.08.024.
6. Nowak DA, Griebl G, Dabitz R, Ochs G. Bilateral anterior opercular (Foix-Chavany-Marie) syndrome. *J Clin Neurosci*. 2010;17(11): 1441–1442. doi:10.1016/j.jocn.2010.02.021.
7. Wu J, Wu Y, Zhang F, Liu H, Hu Y. Where are cortical lesions responsible for opercular syndrome? *Can J Neurol Sci*. 2013;40(1): 97–100. doi:10.1017/S0317167100013044.
8. Milanlioglu A, Aydın MN, Gökgül A, Hamamcı M, Erkuzu MA, Tombul T. Ischemic bilateral opercular syndrome. *Case Rep Med*. 2013;2013:513572. doi:10.1155/2013/513572.
9. Foix C, Chavany JA, Marie J. Diplégie facio-linguo-masticatrice d'origine cortico sous-corticale sans paralysie des membres. *Rev Neurol*. 1926;33:214–219.
10. Starkstein SE, Berthier M, Leiguarda R. Biltateral opercular syndrome and crossed aphemia due to right insular lesion: a clinic-pathological study. *Brain Lang*. 1988;34(2):253–261. doi:10.1016/0093-934X(88)90137-X.
11. Kim JY, Kim JS, Lee JH, Kwon M. Foix-Chavany-Marie syndrome after unilateral stroke. *Eur Neurol*. 2015;74(1–2):84–85. doi:10.1159/000437419.
12. Martino J, de Lucas EM, Ibáñez-Plágaro FJ, Valle-Folgueral JM, Vázquez-Barquero A. Foix-Chavany-Marie syndrome caused by a disconnection between the right pars opercularis of the inferior frontal gyrus and the supplementary motor area. *J Neurosurg*. 2012;117(5): 844–850. doi:10.3171/2012.7.JNS12404.
13. Moragas Garrido M, Cardona Portela P, Martínez-Yélamos S, Rubio Borrego F. Heterogeneidad topográfica del síndrome de Foix-Chavany-Marie [Heterogeneous topography of Foix-Chavany-Marie syndrome]. *Neurologia*. 2007;22(5):333–336.
14. Kobayashi S, Kunimoto M, Takeda K. A case of Foix-Chavany-Marie syndrome and crossed aphasia after right corona radiata infarction with history of left hemispheric infarction. *Rinsho Shinkeigaku*. 1998;38(10–11):910–914.
15. Shoji R, Kono Y, Furuhashi H, Nakano M, Torisu Y. Foix-Chavany-Marie syndrome induced by a unilateral brain abscess. *Intern Med*. 2019;58(4):581–583. doi:10.2169/internalmedicine.1500-18.
16. Torres-Perales AM, Martínez-García FA, Andréu-Reinón ME, et al. Síndrome opercular producido por infarto en un solo opérculo cerebral en un paciente con antecedente de infarto cerebeloso. *Rev Neurol*. 2013;56:495–496.